Denmark, and Iceland. The cases were collected during one and a half years, begin-
ing 1 July 1997.

Method—Clinical variables (including those constituting the EULAR and ILAR criteria) and laboratory findings were registered longitudi-
nally for each patient by paediatricians experienced in paediatric rheumatology. A database was constructed specifically for this purpose. In this study clinical findings and laboratory data from the first 6 months (for certain variables also for the first 12 months) were considered.

Results—322 patients qualified for classification according to the ILAR criteria. Of those 31/322 (10%) children were diagnosed only according to the ILAR criteria. 15% of the patients did not fulfil ILAR criteria for any of the categories and 6% fulfilled criteria for more than one category. As an example of the effect of differ-
cent classification systems in our study, 160/322 were classified as oligoarticular by ILAR criteria, 127/322 as seronegative according to the EULAR criteria. Conclusion—The qualifications for the ILAR criteria are different from those of the EULAR criteria, which makes simple com-
parisons neither possible nor scientifically rele-
ants. Our ability is to study the con-
struct validity of the respective criteria to give us a better idea of the classification and its consequences.

2.5 Juvenile spondyloarthropathies: checking criteria for diagnosis and classification at the start and after 6 months of disease

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Classification of juvenile spondyloarthropa-
thies remains a “critical” point in paediatric rheumatology.
Objective—To validate classification methods in a university hospital treated population by a retrospective cross sectional study.
Methods—Four sets of criteria especially designed for juvenile patients (Garmisch-Partenkirchen-JSA, SEA syndrome, enthesi-

ditis related arthritis, atypical spondarthritides) and two sets of criteria for patients without age specification (ESSG, Amor) were checked in a group of 20 patients at the start of disease and after six months.

Statistics—The sensitivity and specificity were calculated as well as the reproducibility after six months.

Results—The preliminary results show equal sensitivities for all sets except for the SEA syndrome criteria (which was the lowest). Although in some patients a switch to other types of juvenile chronic arthritis occurred, the initial fulfilment of the criteria was highly predictive for the fulfilment after six months.

3 Connective tissue disorders

3.1 Analysis of Raynaud’s phenomenon in an infant and teenage population

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Objective—To analyse the onset and evolu-
tion patterns as well as the prognostic value of Raynaud’s phenomenon (RP) in a cohort of children diagnosed and followed up in our paediatric rheumatology service.

Material and methods—In a prospective study several clinical, analytical, and outcome vari-

ables were collected from a group of patients under the age of 18 years who were admitted to our department because of a history of positive RP.

Results—54 patients (47 female, 7 male) with a mean age of onset of the disease of 13.5 years (range 2–18) were studied. In 31 patients (57%) RP occurred in isolation and in 23 patients (43%) it coexisted with arthritis, sub-
cutaneous nodules and/or hand oedema.

After an average evolution period of 7 years (range 0.9–19) 19% devel-

opled a defined connective tissue disease (CTD) (4 mixed connective tissue diseases, 3 sleroderma, 2 systemic lupus erythemat-
sus, 1 juvenile chronic arthritis), 31 patients (57%) presented some feature of undifferen-
tiated CTD (arthritis, hand oedema, nodules, finger ischaemia, livedo, mucose ulcers, iridocyclitis, positive antinuclear antibodies (ANA), and pathological capillaroscopy pat-
ttern). 13 patients (24%) remain currently as primary RP.

Conclusions—(1) Over 50% (75% in our case) of patients with paediatric onset RP, eventu-
ally develop a CTD or present features consistent with a CTD.
(2) The greater proportion found in our study might be explained by the selection of patients examined.

(3) Arthritis, hand oedema, co-occurrence of ANA and the presence of findings in nail capillaroscopy, have a good prognostic value.

3.2 Severe cardiac disease among children with diffuse cutaneous systemic sclerosis (SSc) and polymyositis: report of 4 cases

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Patients and methods—We retrospectively re-
viewed the charts of 4 girls who met the ACR criteria for diffuse cutaneous SSc and polymyositis, as defined by the presence of proportional muscular weakness and raised secondary serum creatine phosphokinase level. Disease dura-
tion and age on admission ranged from 4 to 12 months and from 7 to 13 years respec-
tively. Dysphagia and oesophageal dysmotil-
ity were present in 3 patients, arthritis in 2, lung restrictive syndrome in 2, and echocar-
diographic abnormalities in 2, including marked non-obstructive cardiomyopathy (NOCM) in 1 child. One patient secondarily developed severe NOCM and bowel disease.

Results—Treatment with cyclosporin, metho-
trexate, and corticosteroids (including pulsed intravenous methylprednisolone in 3 cases) improved the skin thickness score, muscle weakness, arthritis, and lung restrictive syn-
drome. Oesophageal dysmotility and bowel disease did not improve. The 2 patients who had severe NOCM and bowel disease with the ileal combina-
tion treatment died after 19 and 31 months. Severe NOCM developed in a third patient 6 months after treatment onset. Cardiac trans-
plantation was performed and the child is alive and well 3 months after the triple combina-
tion treatment died after 19 and 31 months. Severe NOCM developed in a third patient 6 months after treatment onset. Cardiac trans-
plantation was performed and the child is alive and well 3 months after the triple combina-
tion treatment. One child treated with cyclosporin only is alive after a follow up of 48 months.

Conclusions—As previously described in adults, children with SSc and myositis may be particularly prone to develop severe NOCM.

Earlier and more intensive treatments are probably required. In the case of yet consti-
tuted severe NOCM, heart transplantation should be considered.

3.3 Diagnosis of Sjögren’s syndrome in childhood

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Sjögren’s syndrome is rare in childhood. We have recently seen 5 children with definite and two with possible Sjögren’s syndrome whose investigations highlighted some of the difficulties in diagnosis in the paediatric age group.

Of the children with definite Sjögren’s syndrome one had mixed connective tissue disease (MCTD), one juvenile dermatomy-
sitis, and one systemic lupus erythematosus. Of those with possible Sjögren’s one has MCG. The others have no definite underly-
ing diagnosis.

We used the European classification of Sjögren’s syndrome as modified by Vitali et al 1993 to assess these patients and rapidly became aware of difficulties in using these criteria in children.

Four had symptoms of dry eyes, and six had dry eyes on ophthalmological testing. Three have had symptomatic dry mouths and two have had low salivary flow rates. There are, however, no validated age reference standards for tear and salivary flow rates in childhood.

Sialography, scintigraphy, and histology are used in the classification. Lip biopsy is unacceptable to most children. One patient had a lip biopsy, which proved unhelpful. We have found ultrasound to be well tolerated and to provide information complementary to that obtained by magnetic resonance scanning or sialography. Parotid scintigraphy was per-
formed in two children and thought to be abnormal, but interpretation was hampered by the lack of reference standards in childhood.

Investigation of children with possible Sjö-

gren’s syndrome in our units has highlighted problems with applying the current diagnos-
tic criteria in childhood. There is a need for age appropriate criteria to be validated for use in childhood.


3.4 Life experiences of adolescents living with systemic lupus erythematosus

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Systemic lupus erythematosus (SLE) is a severe chronic disease commonly associated with severe personal, social, and familiar repercussions mainly in adolescents. To get to know the way in which these patients perceive and compare themselves according to the changes and the treatment the disease imposes on their routines, monthly meetings with adolescents with a diagnosis of SLE without central nervous system disease have taken place. Identification of the interests of the group of adolescents was possible through discussion and psychodrama tech-
niques conducted by a doctor and by a social assistant with a degree in psychodrama. The
first concerns referred to by the adolescents were the side effects of steroid treatment (obesity and short stature), the difficulty of the daily use of solar protectors, the limitation on physical activity, the dietary restrictions, how to talk about the disease to friends and relatives, and the constant need for examinations, laboratory tests and testing and, eventually, for invasive procedures (biopsies). The adolescents requested information about the disease itself, its treatment, and the possibility of cure, pregnancy, and contraceptive use. The authors emphasise the importance of the methodology with groups of adolescents with SLE, in which the partner works as a “mirror”—making the adolescents perceive themselves, and as a facilitator—helping them to identify their concerns, anxiety, and expectations. The group was shown to be effective in complementing medical assistance, promoting better adherence to the treatment and a better social and familiar environment through the explanation of several aspects of the disease.

3.5 Neutropenia caused by anti-Ro autoantibodies in neonatal lupus

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Background—Neonatal lupus (NL) provides a situation in which the pathogenicity of autoantibodies can be evaluated. Transplacentally acquired anti-Ro (or SS-A) autoantibodies are found in the sera of babies with congenital heart block or other less common manifestations of NL, such as skin rash, liver disease, or haemocytopenia.

Case report—We report an infant, born to a mother with anti-Ro, who had profound neutropenia. A 30 year old woman with an undifferentiated connective tissue disease gave birth to a healthy girl. Laboratory tests performed on the baby at follow up, 10 weeks after birth, showed normal white blood count, but an absolute neutrophil count of 120/µl.

Causes of congenital and acquired neutropenia include a bone marrow aplasia and were not shown to major abnormalities. IgG anti-granulocyte antibodies were detected in the baby’s serum. Fluorescence activated cell sorting showed binding of mother’s and baby’s serum to intact neutrophils. To determine the specificity of this binding, purified 60 kDa Ro was used as inhibitor. After this treatment, the binding showed binding of mother’s and baby’s serum. Fluorescence activated cell sort showed binding of baby’s serum. To determine the specificity of binding was further studied in sera with 60 kDa Ro.

Various laboratory markers of neonatal cell damage/activation seem to play an important part in rheumatic disease pathogenesis. Among these we have chosen factor VIII related antigens and soluble forms of adhesion molecules ICAM-1 and E-selectin (E-sel) and measured their blood levels in 110 children aged 1-18 years (mean 11.1) with the following diagnoses: polycystic juvenile idiopathic arthritis (JIA) (JIA-poly, n=18), oligoarticular course JIA (JIA-oligo, n=14), juvenile dermatomyositis (JDM), systemic sclerosis (SSc), and overlap syndromes (SSc/JDM, n=11), systemic lupus erythematosus (SLE, n=6), primary vasculitides (VAs, n=90), Henoch-Schönlein purpura (HSP, n=11), acutely ill febrile children (n=10), and healthy controls (n=51). Non-specific inflammatory variables (erythrocyte sedimentation rate, C reactive protein, C3, C4, complement C1q, and protein S) were tested in serum samples of these children.

3.6 Laboratory markers of endothelial damage in the assessment of the activity of childhood rheumatic diseases

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Various laboratory markers of endothelial cell damage/activation seem to play an important part in rheumatic disease pathogenesis. Among these we have chosen factor VIII related antigens and soluble forms of adhesion molecules ICAM-1 and E-selectin (E-sel) and measured their blood levels in 110 children aged 1-18 years (mean 11.1) with the following diagnoses: polycystic juvenile idiopathic arthritis (JIA) (JIA-poly, n=18), oligoarticular course JIA (JIA-oligo, n=14), juvenile dermatomyositis (JDM), systemic sclerosis (SSc), and overlap syndromes (SSc/JDM, n=11), systemic lupus erythematosus (SLE, n=6), primary vasculitides (VAs, n=90), Henoch-Schönlein purpura (HSP, n=11), acutely ill febrile children (n=10), and healthy controls (n=51). Non-specific inflammatory variables (erythrocyte sedimentation rate, C reactive protein, C3, C4, complement C1q, and protein S) were tested in serum samples of these children.

The specificity of binding was further studied in sera with 60 kDa Ro. Such antibodies can directly lower the neutrophil count by binding the cell surface of neutrophils.

3.7 Case report of a patient with catch 22 sequence: haematological and bony abnormalities; the chronic arthritis responds to etanercept

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3.8 Assessment of damage in juvenile onset systemic lupus erythematosus


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Objective—To investigate the prevalence of accumulated damage in patients with juvenile onset SLE and the relation between damage and demographic and clinical variables, overall disease activity, quality of life, and cumulative drug treatments.

Method—A prospective, cross sectional, multinational study was conducted in 218 patients: (a) diagnosis of SLE by ACR criteria; (b) onset age <18 years; (c) disease duration >12 months. Instruments used: SLICC/ACR Damage Index (SDI), SLEDAI, Child Health Questionnaire.

Results—218 patients were enrolled—85 from Italy, 73 from USA, and 60 from Mexico. 86% were female, with a mean onset age of 11.1 years and a mean disease duration of 6 years. 150 (69%) patients had damage. The mean (SD) SDI score was 1.6 (1.8), range 0–12. The organ systems more commonly affected by damage were renal (23%), skin (21%), neuropsychiatric (NPS) (17.5), and ocular (9.7%). Logistic regression analyses...
showed that the presence of damage was positively associated with the SLEDAI score (odds ratio (OR)=1.1, p<0.005), cumulative drug score (OR=1.0, p<0.001), and NPS disease at onset (OR=4.4, p=0.008) and negatively associated with Raynaud phenomenon at onset (OR=0.2, p=0.01).

Conclusions—Our results show that permanent damage is common in pauciarticular onset SLE and is significantly associated with lupus disease activity and cumulative drug treatment. NPS disease at onset is a strong predictor of late damage, whereas the presence of Raynaud phenomenon may exert a protective effect.

3.9 Uncommon causes of liver disease in juvenile systemic lupus erythematosus
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Background—Severe liver disorders are rare in patients with systemic lupus erythematosus (SLE). We report two patients with juvenile SLE who developed uncommon hepatic complications.

Case reports—Patient No 1: A 10 year old girl was admitted with a 3 week history of malar rash, intermittent arthritis, fatigue, and abdominal pain. The diagnosis of SLE was established. During a stay in hospital the girl developed enlargement of the liver, which was tender at palpation. Ultrasonography showed a large thrombosed pancreatic pseudocyst causing the lumen of the inferior vena cava (Budd-Chiari syndrome). Based on the association of vascular thrombosis and positive antiphospholipid antibodies (aCL) and lupus anticoagulant (LA) were detected. The diagnosis of SLE was established. During a hospital stay the girl developed enlargement of the liver, which was tender at palpation. Ultrasonography showed a large thrombosed pancreatic pseudocyst causing the lumen of the inferior vena cava (Budd-Chiari syndrome).

Case No 2: A 9 year old girl, who had had autoimmune thyroiditis since the age of 6 years, was admitted with a 1 week history of fever, fatigue, arthromyalgia, and purpuric rash in the legs. Physical examinations showed hepatosplenomegaly, ascites, and oedema of the legs. Physical examinations showed hepatosplenomegaly, ascites, and oedema of the legs. Laboratory investigations disclosed pancycopenia, liver transaminase increase, hypocalcaemia, low serum complement levels, and positive ANA and anti-DNA antibodies. aCL and LA were absent. Liver ultrasonography showed an enlarged liver with diffuse pseudonodular changes. A percutaneous biopsy showed necrotising arteritis with nodular regenerative hyperplasia of the liver. A diagnosis of SLE with necrotising arteritis of the liver was made, and treatment with high dose prednisone and cyclophosphamide was begun. The patient had no history of jaundice or fever with frequent valvular injuries has a comparatively higher occurrence. In 38% of patients the cause of acute arthritis is unknown.

4.2 High prevalence of childhood chronic arthritis among the Shipibo people of Amazonian Peru
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Few prevalence figures of childhood chronic arthritis exist for developing countries and even fewer for the indigenous population living in a traditional life in the Amazonian part of Peru. Method—A cultural adaptation and linguistic translation of certain key terms of the CR-CHAQ was done. It was tested on children and parents without joint disorders and was with minor alterations well understood. Self administration was not possible, however, as most adults and children cannot read or write. Instead an interpreter gave the Shipibo-CHAQ (SH-CHAQ). The SH-CHAQ was tested on 12 children with possible rheumatic joint disorders, and a close adult. For children below 10 years of age, the SH-CHAQ was applied only to the adult. Results—Cross cultural validity: Most questions had to be adapted to Shipibo lifestyle and traditions. The children gave higher disability scores than the adults. Test-retest was found between the disability index for the first and second application both for children and adults. The correlation for children (Spearman ρ=0.944, p=0.002) was higher than that for the adults (ρ=0.839, p=0.002). Inter-rater score correlation: No correlation was found between the disability index for children and adults at the first application, but the second application showed a good correlation (ρ=0.975, p<0.02). The difference between the first and second application might be due to exaggeration by parents who expected to obtain health care if the child was more severely ill. Conclusion—By carefully analysing the CHAQ and adapting the questions to the Shipibo culture we could successfully apply the new SH-CHAQ with a high test retest and interobserver reliability.

4.4 Remission in juvenile chronic arthritis: a cohort study of 683 consecutive cases
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Evaluation of the prognosis of the juvenile forms of chronic arthritides is difficult because most patients are lost at follow up when they reach adulthood. Our institute is a privileged observatory as it is open to both juvenile and adult patients, so that children do be viewed with caution as the study period was short, the most remote part of the Shipibo area could not be accessed, the concept of medicine is traditional, and long standing reactive arthritis might have been included. This means that the prevalence as well as possibly being underestimated might even have been underestimated. Also among adults of the Shipibo people a very high prevalence, close to 10% has been reported, indicating that the prevalence of chronic arthritis among children also may be high.